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Surgery

Primary Nasosinus Melanoma: A Case Report

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Abstract **Case Report**

Naso-sinusal mucosal melanoma is a highly aggressive but uncommon tumor. It is often diagnosed late, making treatment complex (surgery and/or radiotherapy---), with a generally poor prognosis. The absence of an established etiological factor and a specific clinical appearance make diagnosis difficult at an early stage. This tumor frequently affects women, with a predilection for the 6th decade of life. In this articl, we report a new case of nasosinus melanoma.
Keywords: Nasal cavity, sinus neoplasme, mucosal melanoma, primary cancer, melanoma, head and neck melanoma, unfavorable prognosis, aggressiveness.

INTRODUCTION

Primary naso-sinus mucosal melanoma is a very aggressive and rare malignant tumor (0.6 and 1% of all melanomas).

Diagnosis is difficult due to the lack of specificity of the initial signs. Its aggressive nature and the fact that it is often diagnosed late mean that its prognosis is poor.

The aim of this work and a review of the literature is to specify the clinical, therapeutic and evolutionary characteristics of this malignant tumour of the nasal cavity.

CASE PRESENTATION

63-year-old patient with a Grade 2 Performance Status Scale presenting with a right maxillary sinus process manifested by chronic epistaxis associated with nasal obstruction and cacosmia. Clinical examination revealed a firm, painless mass in the right jugal area, ocular dystopia and a decrease in ipsilateral nostril flow with no limitation of oculomotor movements or diplopia and no cervical adenopathy.

Nasal cavity biopsy with a complementary immunohistochemical study revealed a melanoma facial MRI revealed a locally advanced right maxillary process with right sphenoidal sinus retention.

Rhinocavoscopy showed an ulcerative mass completely obstructing the right nasal cavity, extending beyond the choanae with deviation of the nasal septum to the left associated with bulging of the posterior surface of the cavum explaining the epistaxis.

The pet scan revealed no pathological hypermetabolism, and the therapeutic approach was exclusive immunotherapy.

Figure 1: Photo of patient showing ocular dystopia (right eye above left bicanthal line represented by the blue line)

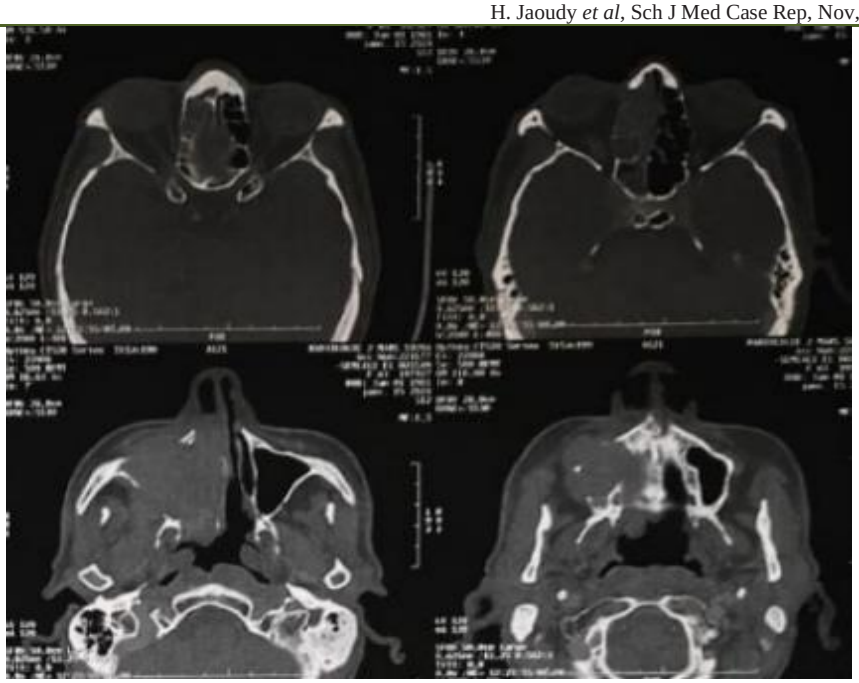

Figure 2: Scannographic images (bone window) showing the process that compresses and blows out the entire wall of the right maxillary sinus, extending to the retrozygomatico-maxillary fat, the homolateral nasal fossa and the choana homolateral. All this is associated with a narrowing of the pterygo-palatine fossa

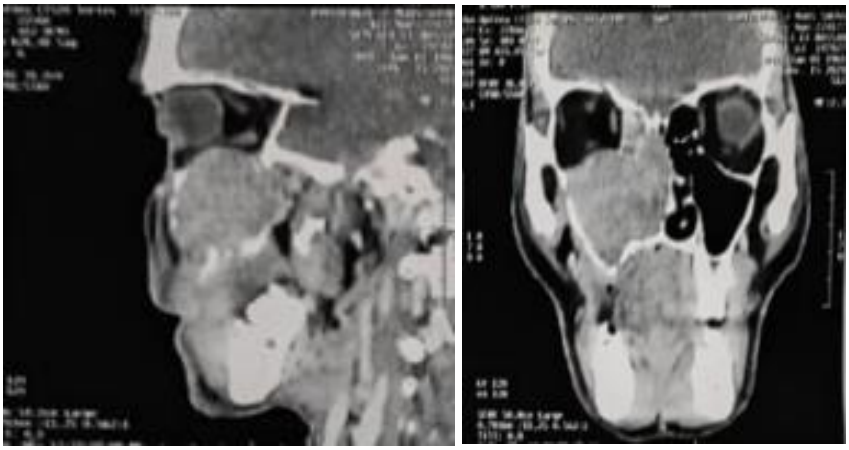

Figure 3: Scannographic images (parenchymal window, (a) sagittal section and (b) frontal section) showing the same lesional process with invasion of the floor of the right orbit, an intimate contact with the eyeball with persistence of a separating fatty border, and nasal refoulement by the nasal extension of the above-mentioned process

DISCUSSION

Naso-sinus mucosal melanoma is rare, but its incidence appears to be increasing. It develops on melanocytes found in the skin and various ectoderm-derived mucous membranes, and less frequently on endoderm-derived mucous membranes such as the nasal mucosa [1-3].

The average age of onset is 65 [4]. The preferred site is the nasal septum, most often the anteroinferior part, followed by the outer wall of the inferior turbinate, then the middle turbinate [2, 3].

Recurrent epistaxis and decreased unilateral nostril flow are the first clinical signs reported by patients [9].

A clinical dermatological examination is essential in this case, as it will enable us to rule out a primary cutaneous melanoma.

Cervical adenopathy is exceptional. Rhinocavoscopy reveals a pigmented or achromic tumour mass, sometimes with melanosis of the neighbouring mucosa [9].

Immunohistochemistry is a valuable diagnostic tool because a pathological study alone is often

inconclusive or may miss the diagnosis, as in our case where an initial biopsy revealed an undifferentiated invasive tumor process.

Surgery is the first-line treatment, with a minimum safety margin of 2 cm. Post-operative radiotherapy is usually performed because of its efficacy for locoregional control [8, 9].

Bacillus Calmette-Guérin (BCG)-based immunotherapy has been used in combination with other therapies (chemotherapy or radiotherapy) to treat isolated cases of nasosinus melanoma. Its efficacy has yet to be assessed, and requires more extensive studies [5, 6].

Recent scientific studies have focused on molecular therapeutics such as growth factors, anti-angiogenic agents and immunomodulating drugs [5, 6].

Unfortunately, the course of the disease is often unfavorable, with recurrence rates as high as 60 to 80%. Survival at 5 years varies from 10 to 47% [6].

Synchronous metastases are rare at the time of diagnosis, but occur frequently during the course of the disease. Clinical prognostic factors are the quality of initial tumour resection, tumour size, recurrence and nodal and distant metastases [4].

Finally, according to some multivariate analysis studies, young age (less than 50 years) and early stage of disease discovery represent good prognostic factors, with an improvement in overall survival [7-9].

CONCLUSION

The prognosis of nasosinusal melanomas can only be improved by early diagnosis, careful locoregional and general workup, and prompt treatment based on wide surgical resection of the tumor.

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